

muscle strength, endurance and flexibility that predispose to chronic or recurrent injury. Because sports-related injuries occur in persons who want to maintain active life-styles, our responsibility is to know enough not only to provide quality medical treatment but also to appreciate and share in an athlete's desire to regain maximal function in a minimal amount of time.

GLEN A. HALVORSON, MD  
Tucson

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## Respirator-Dependent Patients With Duchenne Type Muscular Dystrophy and Amyotrophic Lateral Sclerosis

THERE IS NO CONSENSUS regarding the use of assisted ventilation in the late stage of progressive neurologic diseases. The medical technology required to prolong life with mechanical ventilation has existed and has been refined for more than 50 years. The ethical implications related to quality of life involved in sustaining patients with progressive diseases are a source of continuing concern. Colbert and Schock recently polled the 240 Muscular Dystrophy Association clinics to ascertain physicians' prescribing patterns with respect to this issue. Of 132 responding centers, 24% did not provide respiratory support systems, 33% routinely did prescribe assisted ventilation and 43% provided them only under specific circumstances. The literature of the past decade contains clinical observations and poignant anecdotes advocating both sides of the issue.

The trend among centers where assisted ventilation is used appears to favor methods that do not require tracheostomy. Negative-pressure ventilators used include the cuirass, plastic wrap systems, rocking bed and the Emerson tank (iron lung). Other forms include the Pneumobelt and instructions in glossopharyngeal breathing. Options for positive pressure devices include either intermittent or constant positive pressure ventilation by mouth. Bulbar weakness frequently precludes this method in later stages. Those patients who have reportedly had the most remarkably extended life spans, up to 22 years, have required positive pressure ventilation via tracheostomy. Improved technology, and more lightweight, portable units have made the home use of positive pressure ventilators more practical.

Many patients have been managed with a combination of methods such as positive pressure breathing by mouth in the daytime and a rocking bed at night, or cuirass plus glossopharyngeal breathing. In an intriguing report by Curran, daytime ventilation measured by gas exchange in patients with Duchenne type (pseudohypertrophic) muscular dystrophy was significantly improved by using an iron lung only at night, thus sparing the respiratory muscles. Patients began to use this method of part-time assisted ventilation at the early signs of respiratory failure and were also supported with vigorous chest physiotherapy. The extended life span was accompanied by the requirement of increasingly longer periods on the

ventilator. The possible positive effect of earlier, more aggressive surgical management of scoliosis on respiratory function in this population has yet to be fully examined.

Regardless of the mode of assisted ventilation, decisions about its use are best made electively early in the course of disease rather than later in a crisis of acute respiratory failure. Most articles in the medical literature on this subject imply that the attitudes of physicians and their medical centers weigh heavily in the informed decisions made by patients and families whether or not to choose assisted ventilation. While objective data regarding quality of life are not yet available, physicians are well advised to at least become familiar with the options available to better inform their patients.

ROSS M. HAYS, MD  
KENNETH M. JAFFE, MD  
Seattle

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## Advances in Orthotics and Mobility Devices for Children With Physical Disability

THE PRESCRIPTION of mobility devices for children must take into consideration all aspects of growth and development. During the first three years of life, locomotion and other motor skills are among the primary pathways to learning and socialization. If at all feasible, ambulation is a desirable goal for children even if only for the purpose of exercise.

The traditional bilateral knee-ankle-foot orthosis with pelvic band and hip joint locks allows a tripod gait using upper extremity assistive aids. This gait is unnatural and the energy expended is high. The reciprocation-gait orthosis allows hip motion and provides a more natural and less energy-consuming gait. Also, the orthosis is light, averaging 6 to 7 lbs. The lower extremities, however, must be free of hip and knee flexion contractures with plantigrade feet to accommodate fitting with a reciprocal gait orthosis.

Children with severe limitations in ambulation may find that the vertical wheeler provides a means for safe and rapid mobility. It may be operated either manually with the upper extremities or via a powered mouth-control joystick. An upright posture allows normal eye contact, thus facilitating peer interaction.

After age 1 year, some means of locomotion should be considered for children who cannot move about on their own. The most commonly prescribed mobility device for children with severe physical handicaps is the wheelchair. A variety of wheelchair designs exists, the rear-wheel drive being the most popular. Folding chairs, in particular the light and ultralightweight styles, are available. Originally marketed as competitive sports chairs, they are increasing in popularity. Although they are easily transported, some with snap-off wheels, durability is sacrificed and active children may do better with a standard chair. In growing children, proper seating is important to maintain the correct postural align-